ORIGINAL RESEARCH PAPER
Volume - 12 | Issue - 08 | August - 2023 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

CUTANEOUS CLEAR CELL SARCOMA – A RARE CASE SCENARIO

Oncology

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KEYWORDS

BACKGROUND:
Cutaneous Clear Cell Sarcoma is a rare (1%) soft tissue neoplasm of young adults with melanocytic differentiation(1). Clinically, they present as a slow growing, painless mass over the extremities with a predilection for young females.(2) We report an unusual case of pedunculated and ulcerated clear cell sarcoma arising in the skin of the thigh.

Case Report:
Figure 2- showing Pedicle of polypoidal mass.A 28 years old adult male presented to general surgery OPD with chief complaints of lump over back of right thigh since one year with recent onset ulcer over mass with pain and bleeding for 6 months. He initially noticed a single 1 cm outgrowing pedunculated mass over the back of thigh which rapidly enlarged to current size of 10cm. Initially the skin over the swelling was normal, which later ulcerated with blood and serous discharge for last 6 months and associated with pain. There is no history of trauma, fever. There is no history suggestive of distant metastasis. On examination, there is severe pallor with a single large irregular pedunculated polypoid mass in the posterior aspect of distal right thigh of size 10*8*5 cm extending vertical 20cm distal to left gluteal fold to 5cm proximal to popliteal crease arising from a well-defined pedicle of size 6*2cm, surface is irregular and ulcerated with bleeding points and skin around pedicle is normal. On palpation there is no tenderness and the mass is firm consistency with ulcerated skin over the mass that bleeds on touch. The mass along with pedicle is mobile and there is no palpable inguinal lymph nodes or no signs of metastasis (Figure1&2).

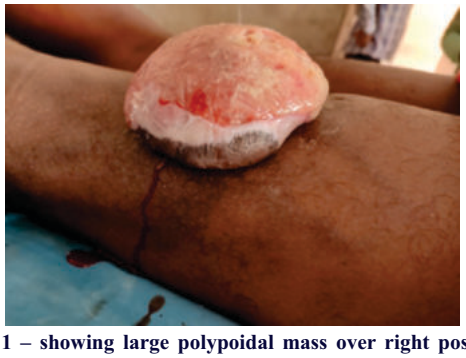

Figure 1 – showing large polypoidal mass over right posterior thigh with ulcerated surface.

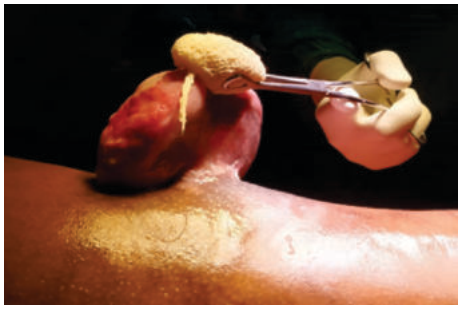

Figure 2- showing Pedicle of polypoidal mass.

On investigation, Plain X-ray of left thigh showed soft tissue shadow with no bony involvement. On ultrasonogram, there was a soft tissue mass arising from subcutaneous plane with no inguinal lymph nodes. His hemoglobin was 7mg/dL, with normal liver profile and negative metastatic work up like chest X-ray PA view and ultrasonogram abdomen.

Pre-operatively his hemoglobin is corrected and the patient taken up under spinal anesthesia for a wide local excision of mass over distal right posterior thigh mass with a 1 cm resection margin of normal skin around pedicle of the mass, and primary closure of wound done.

On cut section, the excised specimen appeared as heterogenous mass with pale white calcifications inside, and specimen sent for histopathology (figure – 3). Post operatively patient recovered well with good healing of sutured wound (Figure – 4).

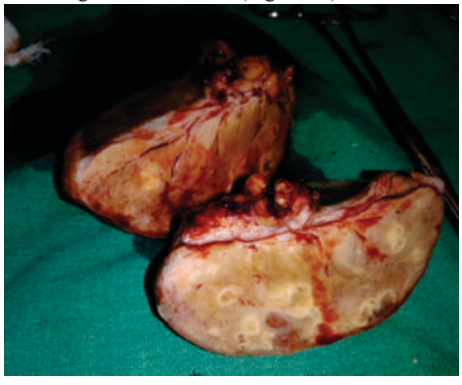

Figure – 3 showing cut section of excised mass.

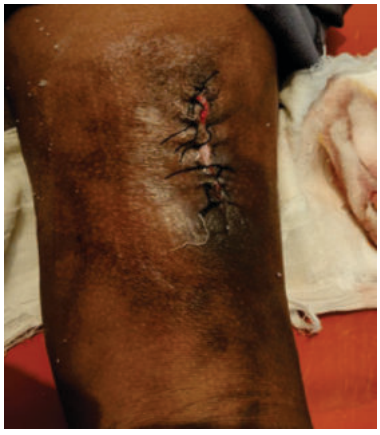

Figure – 4 showing post-operative sutured wound.

The histopathology of the specimen on 10X magnification showed compact nests of cells with clear cytoplasm bordered by a delicate framework of fibro collagenous tissue (Figure – 5). On 40x magnification showed the cells have highly distinctive features consisting of nuclei with vesicular nuclear chromatin pattern and prominent nucleoli. The cytoplasm varies from clear to weakly eosinophilic. IHC staining was positive for anti S-100 and anti HMB-45 antibodies (Figure – 6). The margins are negative. Patient referred to the oncology clinic for further review.

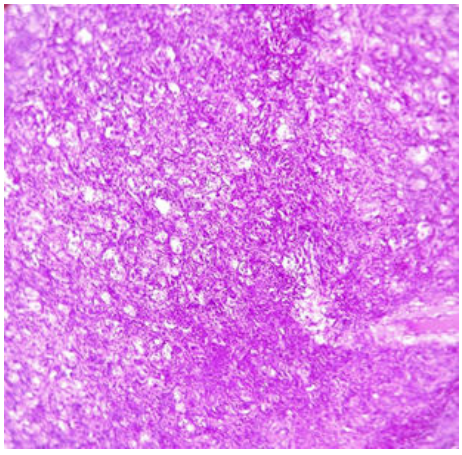

Figure -5 showing 10X magnification of excised specimen.

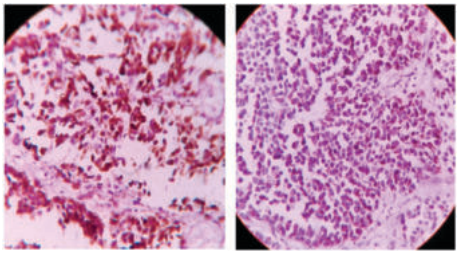

Figure – 6 showing Positive IHC staining with S100 and HMB 45.

DISCUSSION:

Clear cell sarcoma was first described by Dr. Franz Enzinger in 1965 as a rare soft tissue neoplasm, accounting for approximately 1% of all soft tissue sarcoma(3). It is thought to be derived from neural crest cells. It presents as a painful, slow growing tumor occurring commonly in the deep soft tissues, juxtaposed to tendons, fascia or aponeuroses of foot and ankle most frequently(4), although rare cases presenting in the kidney, trunk, penis, gastrointestinal tract, head, and neck have been reported(5). The tumor increases in size which will be followed by metastatic dissemination to lymph nodes and lungs.

Cutaneous clear cell sarcoma mimics the diagnosis of malignant melanoma and hence cutaneous cell sarcoma is called malignant melanoma of soft tissue. On immunohistochemistry, the tumor cells are immune-positive for the common melanocytic markers, namely HMB-45, microphthalmia transcription factor (MITF), S-100 protein, and Melan-A. Ultra-structurally, melanosomes are usually detected. A reciprocal translocation t(12;22)(q13;q12) resulting in a EWSR1/ ATF1 chimeric transcript, identifiable in 70-90% cases, is considered the cytogenetic hallmark of CCS(5). More recent molecular genetic characterization of the 2 diseases (the presence of translocation t[12;22][q13;q12]), resulting in a chimeric gene only in CCS and not in cutaneous malignant melanoma; the presence of activating mutations in the kinase domain of the BRAF gene only in cutaneous malignant melanoma and not in CCS have led to the supposition that CCS and malignant melanoma constitute two separate entities despite their histologic resemblance(6). Distinction of CCS from metastatic MM is important due to the different treatment and prognosis.

Modern treatment of soft tissue lesions requires preoperative workup by X.ray and MRI. MRI probably constitutes the best noninvasive approach in diagnosing a soft tissue mass; it is most helpful in differentiating benign from malignant lesions, and it optimally determines the extent of the lesion. FNA cytology is useful in diagnosing recurrent or metastatic clear cell sarcoma, as reported previously(7).

CCS is a rare and aggressive tumor which makes it difficult to draw conclusions regarding prognostic factors. Tumor size ≥5cm, local recurrence, metastases, presence of microscopic tumor necrosis and DNA-index have been found as poor prognosis factors. Regional lymph nodes or lung metastases have been reported in the patients. Five-year survival rates have been estimated to range from 48-67%.(8) Early detection and local control are of the utmost importance in the management of clear cell sarcoma.(9) Another question to be answered is whether elective lymph node dissection should be performed once the diagnosis of clear cell sarcoma has been established. Lymph node metastases occur in a high percentage of cases, and symptomatic lymph node involvement was a highly unfavorable factor in our series as well as in those reported in the literature. A promising development in this regard is the early detection of occult lymphatic metastases by lymphatic mapping and sentinel lymph node biopsy.(10) Surgery, involving a wide excision of the tumor with sentinel lymph node biopsy, constitutes the mainstay of treatment. Typically, chemotherapy and radiotherapy have not been shown to be beneficial in clear cell sarcoma, but radiotherapy may be used in cases of questionable involvement of the surgical margins. Therefore, it is important to diagnose this entity early and distinguish it from conventional malignant melanoma since further treatments differ significantly(11).

CONCLUSION:

Clear cell sarcoma is a rare, locally aggressive tumor with high recurrence and metastasis. In our case since it's a pedunculated tumor, the tumor did not show signs of nodal or distant metastasis even at stage IIIa. Since this is a rare tumor, there is no extended treatment protocol for this tumor. The effects of chemotherapy and radiotherapy are under evaluation.

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